CEREBRAL SYPHILIS.[1]

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NO class of cases presents such variety of symptoms as exist in syphilis involving the cerebral structure.

Rumpf divides syphilitic diseases of the brain into three classes : first, those involving the cranial bones ; second, new growths involving the brain or its membranes, and, third, specific disease of the cerebral vessels.

In my paper this evening I do not intend to take up the subject of syphilitic tumors or inflammation of the membranes, but shall confine my attention more particularly to disease of the vessels.

Many of the earlier appearances of syphilitic disease of the nervous system, as during the secondary stage, are probably due to simple hypertrophy of the tissues affected.

Virchow describes two conditions as existing pathologically, the one a simple hyperplasia, and the other a heteroplasia ; the latter only, presenting the characteristics of a new growth, distinct and foreign to the part affected, so that for instance, a syphilitic node affecting the cranial bones is of the same distinctive character as a gumma, involving the dura or the pia.

This new growth is the same in the tertiary stage, following, perhaps, thirty years after the original infection, as in the secondary stage, leading one to believe with Rumpf, as opposed to Hutchinson and Ricord, that the virus itself is still active, and that these changes are not due to the effect of a poison which has expended itself, but left its impress on the system.

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[1] Read before section on Practice, of the N. Y. Academy of Medicine, Dec. 18, 1888.

This view is also clinically sustained, as we find many cases of hereditary syphilis transmitted late in the course of the disease, passing through precisely the same class of symptoms as when it is transmitted in the early stages.

The pathological changes in the nervous system in hereditary syphilis do not differ from the acquired form, except that in the former gumma are rare, while in the latter it is the most frequent form of tumor cerebri.

Hutchinson says that cerebral syphilis is rare in the hereditary form, but Dowse and others disagree with him, and find material changes very frequent.

It was long thought that the cranial bones were always the original seat of the cerebral lesion, and although it is frequently so, yet we now know that the dura or pia, or the brain itself may be independently affected.

Meyer considers matting together of the dura and pia with softening of the cortex as almost pathognomonic of syphilis, as in pachy-meningitis, from other causes this rarely occurs.

Characteristic of the pathological changes in specific endarteritis, as distinguished from atheroma according to Heubner, is the fact that in atheroma the large vessels only are affected, and long tracts of the arteries are involved. Occlusion occurs by thrombosis, never by narrowing of the lumen of the vessel ; there is often, at first, even a widening of the lumen of the artery. The process is very slow and consists in simple hypertrophy resulting in calcification, there being no new growth.

In syphilis we find both large and small arteries affected, with a tendency to disease of the small and middle sized vessels. We may indeed have thrombosis, but it is secondary to the narrowing and final occlusion of the vessels from extension of the new growth inwards. Rarely, however, is the endothelical lining of the vessels involved.

The action of the syphilitic virus seems to cause a great increase of cell growth through irritation of the endothelium and commences in the inner coat or intima. When a new growth surrounds the vessels, as in syphiloma or meningitis of the base, the adventitia may be primarily affected.

Arterial disease may occur entirely independent of syphiloma, and is rapid in its course, differing thus from atheroma. According to Dowse the vessels of the pia mater are very often thus affected, leading to softening and atrophy of the cortex and causing thereby a class of symptoms differing from those in which the dura is involved.

The tendency to the formation of new vessels with thin walls so characteristic of all granulation tissue is also present here, so that we have at first a tissue richly supplied with vessels. This of course depends somewhat on the seat of the growth, as in the dura, or between the cranial bones and the dura the tissue is compact, and but feebly supplied with blood, while in the arachnoid and especially at the base the vessels are very numerous.

This rapid cell growth, however, leads to pressure on its vessels, and interference through analmia with its further extension, so that it falls into caseous degeneration, never into suppuration.

This, in a degree, is the same when the disease is limited to the arteries, further extension may be prevented and even retrograde changes may take place, leading to restoration of the lumen of the vessel and disappearance of symptoms. This might readily occur when the cortex is involved, the collateral circulation preventing softening, when, however, the terminal arteries passing to the ganglia and internal capsule are affected, the result may be permanent.

Rumpf places the origin of the endarteritis in the middle coat of the arteries, and insists on its character of granulation tissue similar to that found in all syphilitic growths.

This cell growth may extend inwards, causing occlusion of the vessel and thrombosis. He admits the difficulty of differentiation microscopically from atheroma, but believes that a special bacillus can here be found which is the same for the primary chancre and other later growths.

He shows a drawing of syphilic disease of the vessels, in which the narrowing of the vessels is clearly delineated, corresponding very closely, microscopically, to a section of the basilar artery of a case whose history I will read later.

In the discussion on a paper by Dr. Anderson before the

British Association, on syphilis of the nervous system, Dr. Ross gave the following order of selection in cerebral syphilis :

The cortical area of the middle cerebral artery giving rise to Jacksonian epilepsy ; the cortical area of the anterior cerebral in which somnolence was present and epiliptic seizures infrequent; the subarachnoid space at the base, in which some affection of the cranial nerves ushered in the attack, the disease later extending to the vessels passing into the anterior perforated space, resulting in occlusion of the vessels of the ganglia softening and hemiplegia ; the subarachnoid space at the posterior border of the pons, in which case the disease is ushered in by paralysis of the 6th, 7th and 8th cranial nerves, and a tendency to fall toward the side of the lesion.

In giving a description of the symptoms of cerebral syphilis I shall refer to those of a general rather than a focal character, not taking up tumors as they must be similar in their direct effect to those of any other growth.

I would speak first of the psychical changes present.

These may be sudden in their onset, but more often gradual, extending over months, or even years. We notice a general loss of mental power, the ethical nature seems to suffer, the memory weakens, there is often a feeling of loss of power of performance resembling that of neurasthenia cerebralis; the nature seems changed, there is often developed a tendency to brutality and selfishness, and many acts of intemperance and immorality are committed entirely foreign to the known character of the patient.

The loss of memory, of the association of ideas, and therefore of the judgment, is probably explained pathologically by Clouston's statement that the cortex cells and the associating fibres undergo degenerative changes, such as we know take place in general paresis.

Insomnia, independent of cephalalgia, is one of the early symptoms and always a serious one, or again somnolence or even stupor may occur during the daytime, periodically or continuously.

This is not to be confounded with the coma of an apoplectic seizure.

Cephalalgia is perhaps one of the most frequent of all the early symptoms and indicates an affection of the cranial bones or of the dura mater, it may be worse at night, and is always accompanied by a feeling of mental weakness and confusion. Cephalalgia is not always present, indeed is usually absent when the lesion is in the arteries or in the brain itself; the psychical confusion, however, always exists.

The cranial nerves are frequently affected, especially the 3d, 6th, 5th, and the optic. We may have these nerves bilaterally or unilaterally affected, thus ptosis of one side alone, or again the 5th nerve of one side only involved.

Bulbar paralysis, according to Buzzard, may occur, although I believe it is rare. Hutchinson says the 7th alone is rarely affected, and that he has never seen a case, although others (Jackson) have reported them. He is equally strenuous on the diagnostic value of an affection of the 5th as indicating syphilis.

In tumors or meningitis of the vertex these nerves necessarily escape, while where the base is involved many, including the facial, are affected. It is however in disease of the vessels unassociated with tumors or meningitis, we often find the single paralysis of the cranial nerves.

Epileptic seizures are often present, and, as the inflammation is usually diffuse rather than circumscribed, they are more often general than focal. This is seen in cases of lepto-meningitis, with escape of the dura and cranial bones. Following these attacks the coma is apt to be profound, with often a maniacal condition and paralysis transient in character.

In regard to another of the early symptoms, I would direct especial attention to the eyes.

In the secondary stage of syphilis, as we know, the iris and cornea are frequently involved, and usually symmetrically, and in doubtful cases it is well to bear this in mind.

I would especially refer to choroiditis and retinitis as indicative of an already existing affection of the cerebral vessels. A cloudiness of the disc, appearances of atrophic spots in the fundus, traces of a post retinitis or choroiditis, gummatous enlargements involving the terminal vessels, aid in a positive diagnosis.

F. Ostwalt, in an excellent paper in the Berl. Kl. Wochenschrift, No. 45, 1888, dwells particularly on these points, and draws a very important conclusion from them, that they are secondary to changes in the carotid artery, the ophthalmic artery giving symptoms much earlier than the cerebral vessel with its more complete collateral circulation.

As a diagnostic point, an interference with vision in any part of the field, or a general loss of acuity of vision in the young, should lead to the suspicion of cerebral syphilis. The writer relates a case where two or three such attacks preceded by several years a fatal hemiplegia.

Hutchinson notes that in hereditary syphilis, often as late as the tenth to the sixteenth year, corneal affections, usually symmetrical, may occur; and that often, when no signs of the disease have shown themselves during the first two years, later, mental weakness may develop, or melancholia, mania, or dementia. He also mentions a symptom which I have not found generally referred to—the appearance at this age of deafness without local cause, for which there seems to be no cure.

The early recognition of symptoms is of great importance, as later treatment cannot be of much avail.

Transitory paralysis and paralysis of parts widely separated are strongly diagnostic of specific disease.

An hemiplegia coming on without loss of consciousness, in which the patient can relate the commencing numbness and tingling, and final paralysis as it spreads from the arm to the leg, especially if the condition has been preceded by irritability, loss of memory, cephalalgia, confusion, somnolence, in a person under forty, is probably specific.

Many, if not precisely this class of symptoms occur in the aged, due to thrombosis.

It is not unusual in these cases to observe a tendency to somnolence or even stupor, passing into coma and paralysis, the latter often being only temporary. In these cases, however, we have the advanced age, the atheromatous arteries, or some chronic kidney disease pointing to the cause.

In examining some fifteen or twenty cases of hemiplegia from various causes, I found, out of six of specific origin, five affecting the left side; and while this could not be taken in any sense as the proportion, yet it suggests one fact, that hemiplegia in syphilis is more often on the left side than in other cases; in other words, it may as often affect the vessels of one of the hemisphers as of the other.

In the discussion on Dr. Anderson's paper, before alluded to, Clouston gives several principal forms of brain syphilis. First, where we have a short delirium during the secondary stage; second, acute delirious mania during rapid growth of a syphiloma in the cortex, in the region of the vertex anteriorly, with rapid progressing softening, the patient dying within three weeks of the incidence of the disease; and third, the most common form he ascribes to syphilitic endarteritis, the mental symptoms being immorality, sudden impulsiveness, insane suspicions, and gradual dementia accompanied by convulsions and paralysis.

Buzzard says that multiple symptoms are diagnostic of specific disease of the nervous system, the occurrence of a lesion involving two parts of the cerebro-spinal system being rare except in syphilis.

Transitory paralysis may suggest hysteria, as in a case reported by Heubner, in which extensive specific affection of the arteries and membranes was found post-mortem.

Cerebral syphilis may also be confounded with neurasthenia cerebralis, the mental inactivity, depression, and apathy rendering it very difficult of diagnosis, especially in those cases where headache is absent.

It is especially in these cases that a careful inquiry into the previous history becomes necessary, and an examination of the field of vision. The presence of signs of a previous iritis or choroiditis, also of any signs of thickening or of nodes affecting the skull, clavicle, sternum, or tibia, of old scars, especially if pigmented or concentric, of atrophy or hypertrophy of the testicle, of lesions of the tongue, etc., is indicative of specific disease.

Epileptic seizures after twenty years of age should lead to the suspicion of syphilitic origin when injury or other causes can be excluded. As in idiopathic epilepsy, the cases are most common between ten and twenty years.

According to Hughlings Jackson, in these cases we may have only a slight contraction of the fingers or the face without loss of consciousness, or, again, very rapid succession of fits with intervals of deep coma.

Rumpf lays great stress on the pupil changes as a diagnostic point in cases where no focal lesion is present, an inequality of the pupils, or the more active response to light of one than the other.

The Argyl-Robertson pupil, one of the earliest symptoms in tabes, he considers also an initial symptom in cerebral syphilis, and quotes Moeli and Oppenheim as agreeing with him. This is of especial importance in a differential diagnosis from neurasthenia cerebralis, and the writer relates a case which recovered under specific treatment, in which this was the only symptom pointing to a syphilitic origin.

I consider it of great value, and have tested it in several cases which subsequent treatment and examination proved to be specific.

In conclusion I would detail briefly a few cases that have come under my care in private and hospital practice which more or less illustrate many of the symptoms and pathological changes given above.

CASE I.—A. M., colored, female, æt. 35; unable to obtain family history. Patient married at 16; sterile; specific history. Her sister says that the patient was a temperate and hard-working woman up to the summer of 1887, when she became changed in character, and frequently came to this city from her home in Brooklyn, and became intoxicated and went with lewd company. Patient had never complained of headache or trouble with sight or hearing, so far as she knew. Patient was admitted to the workhouse in

September, 1887, for disorderly conduct. The house physician reports that she was irritable, indecent in speech, confused, and did not seem to hear or understand what was said to her. Soon after admission had an epileptic seizure, general in character, followed by deep coma and mania in which restraint was necessary.

Following attack patient was blind, groping about and knocking into objects in her way. Her condition remained the same until January, 1888, similar attacks occurring every three weeks. I saw patient the day following the last seizure—condition partly comatose; position in bed one of flexion; she could be roused, but would obstinately cover herself with the bed clothing, and laugh in a silly manner; reflexes normal, and as far as could be ascertained no loss of response to touch, pain or temperature tests. There was a slight persistent tremor of right side, but no paralysis. Patient died two days later in deep coma, following a maniacal attack.

Post-mortem examination showed the following :

Skull normal, no thickenings or exostoses; dura mater not adherent or thickened; slight lepto-meningitis over convexity of both hemispheres parallel to longitudinal sinus. Extensive softening and atrophy involving bilaterally and symmetrically the parietal lobes posteriorly to the motor area; the left occipital lobe much atrophied.

The vessels supplying these parts, *i. e.*, the branches of the middle cerebral and posterior cerebral were occluded or narrowed and the walls thickened.

Microscopical sections of the basilar and middle cerebral made by Dr. J. S. Thacher, showed thickening of the walls of the vessels, the intima and middle being principally involved and the lumen encroached upon.

His report is as follows :

"The intima is in spots very markedly and irregularly thickened, appearing to consist in such places of new connective tissue containing some blood filled spaces.

"Some of the deeper portions of such thickened patches showed clusters of large pale staining, coarsely granular cells which I took to be fatty degenerated cells, others of the deeper portions showed masses in which no structure could be made out.

"The media was in circumscribed places much thinned, making a kind of hole filled by thickened intina.

"Beyond this I detected no change in the media and none in the adventitia."

I present these sections along with another from a case of known specific history, in which the extension of the growth into the lumen of the vessel is very beautifully seen.

CASE II.—A. B., merchant, æt. 42. Patient enjoyed good health up to two years before coming to me. Patient complained of cephalalgia diffuse in character, vertigo, depression, incapability of sustained mental work, and insomnia, in short, many of the symptoms of so-called neurasthenia.

Patient denied syphilis, and with the exception of some tenderness on percussion over the vertex I could detect no signs of it. Under moderate doses of potass iodide complete recovery.

CASE III.—C. D., female, æt. 42. Patient had complained of trigeminal neuralgia for weeks, and had received the usual remedies. On examination I found the pupils unequal and slow in response to light, although responsive to accommodation; patient also mentally dull.

Suspecting syphilis large doses of the iodide were ordered with complete recovery. A more careful investigation into the previous history also sustained the diagnosis.

CASE IV.—E. D., female, æt. 29. Saw patient for first time while affected with chorea and iritis of both eyes. Syphilis was denied, but I found on examination enlarged glands on the neck and several scars over the body. Patient excessively irritable, refusing food or medicine, remaining in bed in the daytime only to disturb the ward at night with her hallucinations. Later knowledge showed that she had been in the insane asylum for one and a half years.

Under potass iodide and calomel—small doses—chorea and iritis much improved.

In regard to treatment the iodide must be relied upon. Large doses, up to 300 or 400 grains daily if necessary. Often when this fails, mercury, in conjunction with it, either by inunction or the hypodermic method, seems to be very effective. In all these cases the nourishment should be carefully watched and carried to as high a point as possible.